The Royal College of Surgeons of England

doi 10.1308/147870806X129241

Online Case Report

Scoliosis development in identical twins after intercostal thoracotomy for pulmonary artery sling correction

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We present a report on two monozygotic female twins who underwent a left-sided intercostal thoracotomy in the second month of life for pulmonary artery sling correction. Twenty-four years later, in adulthood, the identical twins had both developed right-sided thoracic scoliosis. No previous accounts of scoliosis development after intercostal thoracotomy for pulmonary artery sling correction have been reported.

Keywords: Scoliosis – Pulmonary artery sling – Thoracotomy

Pulmonary artery sling is a rare congenital disorder in which the left pulmonary artery arises from the posterior of the right pulmonary artery. Death can occur in the first few months of life due to tracheal and right bronchus compression. Surgical correction at this time can minimise the high mortality rate associated with medical management. This requires an intercostal thoracotomy or midline sternotomy.

Case report

Two female monozygotic twins were diagnosed with pulmonary artery sling anomaly in the first few weeks of life at another institution. Surgical correction was identical and undertaken at 2 months of age by the same cardiothoracic surgeon, one immediately after the other. The approach consisted of a straight lateral thoracotomy at the left 4th intercostal space with thoracic access gained using a rib spreader. No rib or other abnormalities were identified at the time of surgery. Neither twin suffered any immediate complications after their surgery. As discussed previously, both twins had recovered well by the 2-year postoperative stage.[1]

At the age of 24 years, twin 1 presented to our institution with increasing back pain over the preceding year and a progressing thoracic scoliosis. Since 16 years of age, she had experienced mild thoracic discomfort at which time her family noticed a mild scoliosis. After informed consent, examination revealed twin 1 had developed a right-sided thoracic scoliosis with level and symmetrical shoulders and pelvis. A left-sided intercostal thoracic scar was visible. Radiographs illustrated a 55° right-sided (convex to the right) thoracic scoliosis with apex at the T8 vertebral body, a lumbar compensatory curve and 12 degree rib hump (Fig. 1). Twin 2 was also noted to have a right-sided thoracic scoliosis and a left-sided thoracic scar at the same level. However, her thoracic scoliosis measured only 29° without rotation and lumbar scoliosis (Fig. 1). She had also experienced mild thoracic discomfort and noticed a spinal curvature at 16 years of age. No rib humps, neurological abnormalities or other problems were detected in either twin. Whole spine MRIs identified no spinal or neural tube anomalies. No congenital spinal problems, such as hemivertebrae, were detected.

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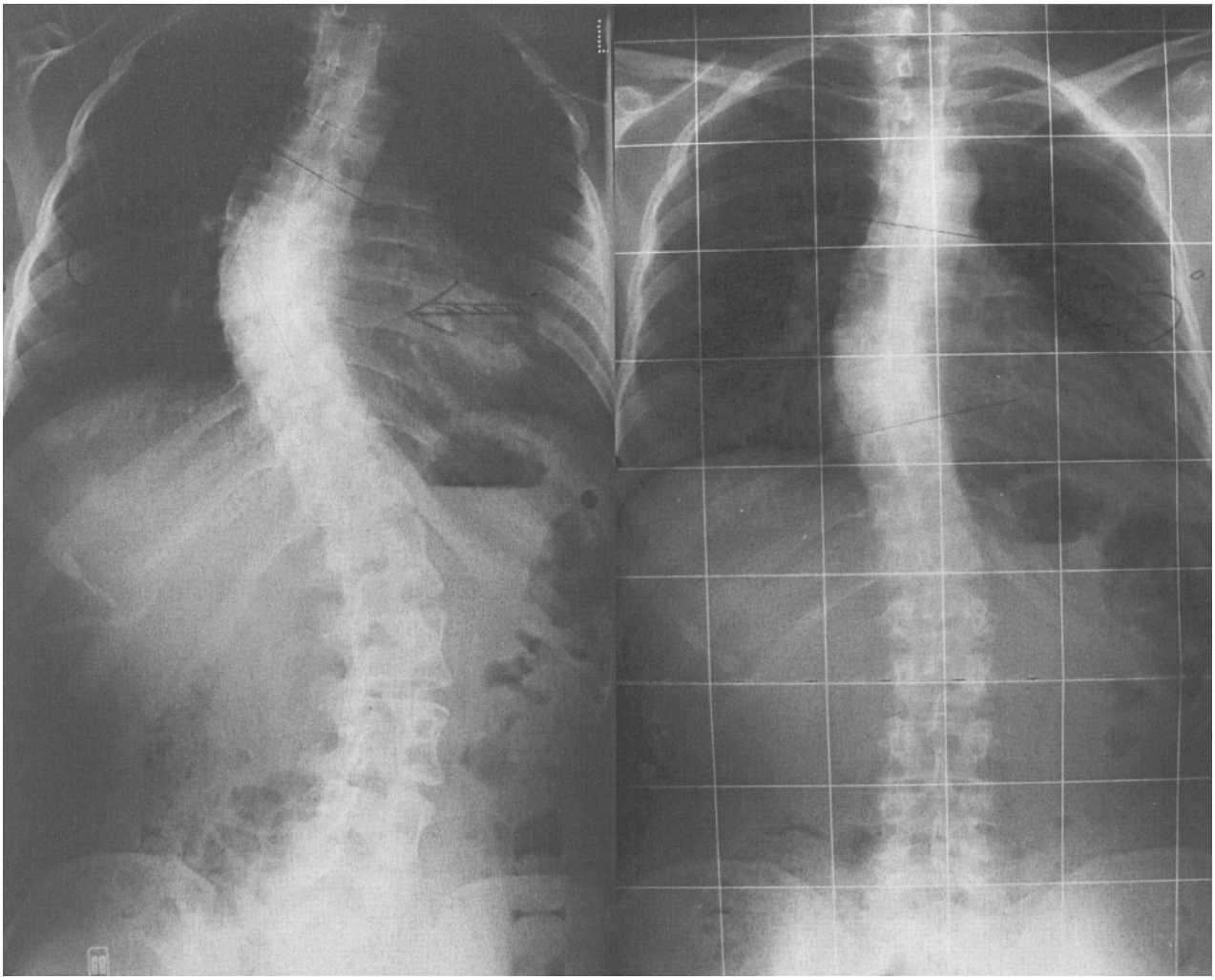

Figure 1 Radiographs indicating 55° scoliosis in twin 1 (left) and 29° scoliosis in twin 2 (right).

Discussion

Survival of symptomatic infants with pulmonary artery sling is unlikely without early surgical intervention. Surgical repair involves division of the anomalous left pulmonary artery and re-anastomosis to the main pulmonary artery anterior to the trachea. This can be performed using a left thoracotomy or a midline sternotomy. The latter provides better exposure of the pulmonary artery, eases cardiac bypass and allows associated tracheobronchial abnormalities to be corrected, which cannot be performed easily using a left thoracotomy approach. This technique is associated with a low operative mortality and excellent left pulmonary artery long-term patency.[2]

Early research attributed scoliosis development in congenital heart disease to an increased heart size. More recently, treatment of congenital heart disease using thoracotomy has been implicated. Postoperative scoliosis development is associated with left-sided thoracotomy for paediatric congenital heart disease. A high-risk period for scoliosis development after aortic co-arctation correction commences 3 years' postoperatively.[3] Scoliosis also occurs after thoracotomy for other thoracic conditions such as tracheo-oesophageal fistulae and oesophageal atresia, in which the development of rib fusion has been associated. Scoliosis can result in cardiorespiratory compromise, back pain, impaired mobility and deformity. Early surgical correction can limit these complications. However, when greater curvatures are treated late, more complicated surgical procedures are often required.

Research indicates a greater frequency for convex curvatures toward the side of incision after lateral thoracotomy for congenital heart disorders, pulmonary malformations and oesophageal atresia.[4] Explanations for the direction of scoliosis development are conflicting but include rib tethering. No definite cause for the variability in direction has been identified. The relationship between site of the curvature, its magnitude and the site of thoracotomy appears complex and does not fit a set pattern.

Several non-surgical associations have also been identified for scoliosis development. Genetic factors play a pivotal role in determining scoliosis development after birth. Kesling *et al*.[5] detected a significantly higher risk of scoliosis development in monozygotic twins compared to dizygotic twins and a statistically high correlation between curve severity in monozygotic twins not seen in dizygotic twins. Familial studies have indicated a polygenic method of inheritance. Environmental factors *in utero* and after birth have also been implicated. Congenital heart conditions have been more frequently associated with congenital osteogenic scoliosis. However, no congenital spinal problems were identified in the presented twins.

Conclusions

The monozygotic twins presented here show that scoliosis development may occur later in life in patients who have undergone paediatric surgical pulmonary artery sling correction. The likely cause is adolescent idiopathic scoliosis. However, multiple factors including surgical approach, genetics and environment may have influenced development. Specialists caring for such patients should be alert for the development of scoliosis in childhood and early adulthood. The authors advocate similarly treated children should be regularly reviewed until cessation of growth. When concerns of scoliosis development arise, review by orthopaedic spinal specialists can aid early detection of progression and institution of treatment.

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